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A CASE OF PROGRESSIVELY DEVELOPING HEMIPLEGIA, LATER BECOMING TRIPLEGIA, RESULTING FROM PRIMARY DEGENERATION OF THE PYRAMIDAL TRACTS.[1]

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In this contribution the clinical and pathological details of a case of progressively developing hemiplegia, which later became triplegia, due to primary degeneration of the pyramidal tracts, will be presented, with a review of the literature of similar clinical cases. A summary of the literature of primary bilateral sclerosis will also be given.

J. S., white, was admitted to the Philadelphia Hospital August 9, 1897, where he died January 14, 1903. He was fifty-four years old when admitted. With the exception of a short time during which he was treated in the Men's Medical Ward for diarrhea or dysentery, he was during this period of six years a patient in the Men's Nervous Ward.

Omitting some unessential points, his history as traced in the hospital records is as follows: The family history obtained has no etiological value. He denied venereal disease. He was a heavy drinker and also used tobacco. He had had rheumatism six years before admission to the hospital. The earliest note referring to any nervous symptoms was made August 9, 1897, when a statement was recorded that two years before admission he had been paralyzed on the right side. It is probable that at this time (during 1890-1895) his loss of power first began to show itself.

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[1] Read at the meeting of the American Neurological Association, May 12, 13 and 14, 1903.

From the William Pepper Clinical Laboratory, Phœbe A. Hearst Foundation.

On September 28, 1897, the record stated that right hemiplegia (more correctly hemiparesis) was present, that the biceps-jerk and knee-jerk were increased, and that the plantar reflex was absent. Ankle clonus also was absent.

On September 24, 1899, it was noted that he could use his right leg for various willed movements, but that it showed loss of power as compared with the left; also that there was some loss of power in the right arm as compared with the left. His grip measured by the dynamometer indicated fifty-five for the right and seventy-five for the left. Speech was a little thick. The knee-jerk on the right was much exaggerated, and the quadriceps-jerk and gastrocnemius-jerk were also increased. Both ankle-clonus and patellar clonus of moderate degree were present on the right. No note was made at this time as to the Babinski phenomenon. Biceps-jerk, triceps-jerk and all muscle jerks in the right upper extremity were increased. The tendon and muscle responses on the left were all present but not exaggerated as on the right, and patellar clonus and ankle clonus were absent on the left. Sensation for touch and pain was retained all over the body, and the stereognostic perception was undisturbed. The irides reacted to light and in accommodation, and other examinations for nervous phenomena gave negative results.

On May 17, 1901, the patient complained that the right leg was weaker than it had been one year previous. The reflexes and other symptoms were much as before recorded.

Although the case was seen from time to time no further notes as to his nervous symptoms were made until December 29, 1902, at which time it was recorded that the movements of the face and tongue were normal; the upper limbs could be moved voluntarily and fully in all parts; the grasp of the right hand was feeble, of the left normal; voluntary power in the right lower limb was very slight; in the left lower limb it was a little better than in the right but much impaired. The right upper limb and both lower limbs were spastic, but contractures and wasting were not present. The biceps, triceps and wrist reflexes were much exaggerated in the right upper extremity, and in the left they were a little more prompt than normal. Patellar and Achilles reflexes were exaggerated on both sides, but more so on the right; patellar clonus was present on the right but not on the left; ankle clonus was elicited on each side. The Babinski reflex was present but not very prominent on the right; it was absent on the left. Sensation to touch and pain was everywhere normal.

This man suffered at times from severe intestinal and cardiovascular symptoms. On August 6, 1897, he began to have severe pain in the abdomen with diarrhea, with numerous stools accompanied by straining and pain. He was treated for these symptoms in the medical wards. The heart sounds at this time were feeble and accompanied by a low systolic murmur at the right base. The sounds were feeble on the left also, and were best at the ensiform cartilage. On December 11, 1898, the records show that he had a soft blowing systolic murmur at the second aortic interspace. May 31, 1899, the patient complained of pain in the chest most marked over the precordia. A soft double systolic murmur was heard above the second cartilage. The heart's action was weak and irregular. A few unimportant notes, chiefly negative, were made in September, 1899. The presence of the cardiac murmurs above described was reiterated. On January 17, 1902, the patient complained of constant and severe pain over the precordial region; the heart sounds were very feeble. The pulse on the left side was imperceptible, owing to the atheromatous condition of the arteries and to the cardiac weakness. The pulse on the right side was perceptible but weak, and the radial arteries on both sides were hard and rigid. The patient suffered at this time and later from cardiac weakness and anginal pains for which he was treated with various remedies.

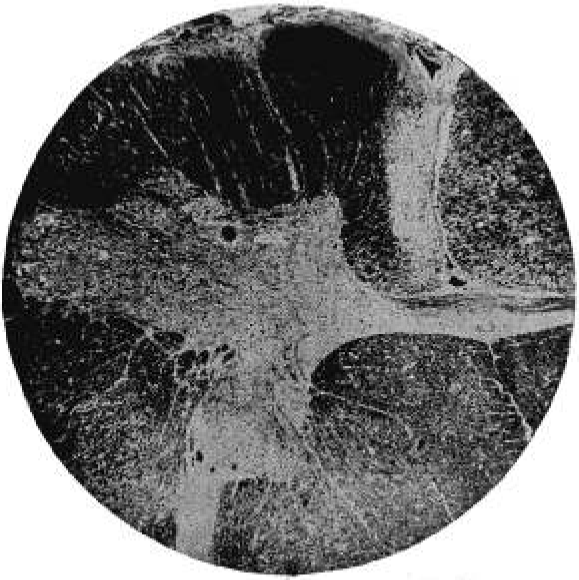
About January 1, 1903, he began to show signs of great weakness and rapid failure. He had at times attacks of vomiting. He lost flesh, his pulse became weak and intermittent, and the right lung became edematous. He died January 14, 1903, his death being preceded by a slight convulsion.

The necropsy showed a serious pathological condition of the vessels and many of the viscera: chronic pleurisy and tuberculosis with cavity formation; endocarditis, contracted kidneys and chronic marked atheroma. The heart muscles were soft and pale, the valves distorted, thickened and calcareous, the aortic extensively atheromatous. Both kidneys were very firm, dark in color, with granular surface and irregular cortex. The stomach was decidedly contracted and the seat of chronic catarrh. Beginning at the junction of the transverse and descending aorta was a unilateral dilatation of this vessel, which increased in diameter to about 4 cm. This structure was atheromatous and contained a flattened clot. The brain and cord were removed for microscopical examination, of which the following is a record.

The degeneration in the lumbar region was very distinct by the Weigert hematoxylin stain in the crossed pyramidal tracts, and more intense in the right than in the left. The direct pyramidal tracts by this stain did not appear degenerated at this level of the cord. The nerve cells of the anterior horns were not distinctly altered, they were numerous and some were much pigmented, which, considering the man's age, was not remarkable. The chromophilic elements were well formed and the nuclei central. It may be said that the nerve cells of the anterior horns of the lumbar region had the usual appearance in persons of the same age. The anterior roots on each side in the lumbar region were normal.

At the eighth cervical segment the degeneration of the right crossed and left direct pyramidal tracts was very distinct by the Weigert hematoxylin stain. The left crossed and right direct pyramidal tracts were much less degenerated. The nerve cells of the anterior horns of this region may be said to have been normal, although in examining a number of sections a few cells were found that were not normal; the nerve cells in general were remarkable for their excellent state of preservation.

Sections from the lower cervical region stained by the Marchi method showed no recent degeneration in the right crossed and left direct pyramidal tracts. The left crossed and the right direct pyramidal tracts contained numerous small black dots, and those of the crossed tract were much smaller and less numerous than those of the direct tract. At the outer border of the right direct



Long-standing degeneration of the left direct pyramidal tract, recent degeneration of the right direct and of the left crossed pyramidal tracts. The dots in the crossed tract are not so large as those in the direct tract (Method of Marchi).

pyramidal tract the dots were smaller than in the median and inner portions of this tract.

The anterior roots in the lower cervical region were normal. There was no distinct meningitis; here and there a slight accumulation of round cells was found in the pia.

Both pyramidal tracts were much degenerated in the oblongata and pons, but the left tract was much more degenerated than the right. The right pyramidal tract was much degenerated in the oblongata and pons by the Marchi method, while the left showed no recent degeneration by this method. About the middle of the pons, the left pyramidal tract contained a large proportion of normal nerve fibers, but a part of it was much degenerated. No degeneration could be detected in the foot of the right or of the left cerebral peduncle by the Weigert hematoxylin method, but degeneration by the Marchi method could be found in the middle portion of the foot of the right cerebral peduncle and also where the foot of the right cerebral peduncle becomes the lower part of the posterior limb of the internal capsule; the degeneration however was slight. No degeneration could be seen by the Marchi method in the foot of the left cerebral peduncle.

Sections of the left paracentral lobule contained Betz cells that appeared normal. They were not numerous, but this does not mean that there had been a loss of the cells, as the number of these cells seems to vary greatly in different cases.

Cellular infiltration of moderate intensity was found in the pia about the cerebral peduncles and pons, but it seems hardly probable that this could explain the long-standing degeneration of the pyramidal tract from the left cerebral hemisphere, and for this reason we have spoken of the degeneration of the pyramidal tracts as primary. No focal lesion could be detected anywhere.

Summarized, this case was one in which hemiplegia gradually developed on the right side, the lower extremity being more markedly and probably earlier affected than the upper, the case therefore at first belonging to the clinical type of unilateral progressive ascending paralysis. After several years the left lower extremity also became paralyzed, but not to the same extent as the right. The reflexes were all markedly exaggerated, the Babinski response being present. Sensory symptoms were absent. Microscopical examination showed intense and long-standing degeneration of the right crossed and the left direct pyramidal tracts, the degeneration extending into the pons but not into the left cerebral peduncle; also comparatively recent degeneration of the left crossed and the right direct pyramidal tracts, traced by the method of Marchi into the lower part of the right internal capsule. No lesions, degenerative or focal, were found elsewhere in the brain or spinal cord; the case, therefore, was one of primary degeneration of the motor tracts, much greater and older in the right crossed and left direct pyramidal tracts. The case may be regarded as a corroboration of the clinical type described by Mills in the JOURNAL OF NERVOUS AND MENTAL DISEASE for April, 1900.

In the JOURNAL OF NERVOUS AND MENTAL DISEASE for April, 1900, in a paper entitled "A Case of Unilateral Progressive Ascending Paralysis, Probably Representing a New Form of Degenerative Disease," which was presented to the Philadelphia Neurological Society on December 18, 1899, an interesting case is detailed, reference being made to another somewhat similar case previously observed. The patient, a man fifty-two years old, about two years before coming under observation began to show signs of weakness in the right lower extremity, as indicated by his method of walking. The paresis came on slowly and insidiously. Weakness in the arm appeared for the first time eighteen months after the weakness was noticed in the lower extremity. This weakness soon became more and more evident, and was accompanied by the tendency to carry the arm raised against the body and flexed at the elbow. When first examined the paresis in the upper extremity, although easily determined, had not nearly reached the degree of impairment observed in the leg. The right side of the face was also slightly but undoubtedly paretic. The patient had had at one period some hyperesthesia in the right lumbar region and in the lower extremity, and later in the right upper extremity. Herpes appeared in the lumbar and lumbosacral regions, lasting for a short time. Wasting was distinct in the right lower extremity, the measurements showing a difference of one and a half inches for the thigh, and five eighths of an inch for the calves. The various movements of the right leg were distinctly weaker than those of the left, but were nowhere absolutely abolished. Similarly all the movements of the right arm were distinctly impaired, but were nowhere absolutely lost. The dynamometer showed 180 for the right and 160 for the left. Faradic contractility was retained. The affected limbs were not spastic nor contractured. Careful examination showed retention of all forms of sensation. The tendon and muscle phenomena on the right side were all somewhat exaggerated. Knee-jerk was plus on the left side, but was considerably more exaggerated on the right. Patellar clonus was present on the right but not on the left, and the right side showed a slight ankle clonus, which was absent on the left. The plantar reflex was normal on the left; but on the right, while the Babinski reflex was not present, the normal response was distinctly less marked than on the left. It might be described as between normal plantar flexion of the toes and the dorsal flexion of the Babinski reflex.

This case has recently been made the subject of reinvestigation by Dr. Hugh T. Patrick, who inclines to the opinion that the case is one of paralysis agitans without tremor, the disease up to the present time having attacked only one side of the body. His report of the case was presented at the meeting of the American Neurological Association, and will be found in its Proceedings, with the discussion of the case by Dr. Mills. At the time when the patient was examined by Dr. Mills, among the reasons for believing that the case was not paralysis agitans were the absence of spontaneous tremor; the presence of markedly exaggerated reflexes on the affected side, even including ankle clonus and patellar clonus; the existence of decided wasting on one side, especially of the lower extremity, and the absence of the facies of paralysis agitans and of a fixed position of the body and head.

According to Sir William Gowers, both primary lateral sclerosis and paralysis agitans are probably abiotrophies (*Lancet,* April 12, 1902), and it may be that in the case under consideration the two abiotic diseases are conjoined.

In the same paper in which this case is detailed a second case is reported. This patient was under the care of Dr. Mills seventeen or eighteen years ago, and had previously been a patient of the late Dr. E. C. Seguin of New York, who believed that there was a cerebral lesion causing changes in the crossed pyramidal fasciculus analogous to lateral sclerosis. The view taken by Mills was that the changes in the crossed pyramidal tract were of the nature of a lateral sclerosis, but were primary and not secondary to any cerebral lesion. The patient was a woman fortythree years old, who first noticed weakness in her leg while pregnant with her last child, three years before coming under observation. The left arm became paretic a few months after the left leg. The reflexes were much increased on the affected side. The patient remained under observation for many months, the paresis of the leg and arm slowly increasing. Neither arm nor leg was contractured, and sensibility was preserved. The patient complained at times of pains like neuralgias in the limbs, and of nervous twitching in the leg and arm. This patient was alive three or four years ago, and had become entirely unable to walk, but just how she was affected was not learned.

In the paper reporting these cases the differential diagnosis from an unusual form of unilateral disseminated sclerosis, from unilateral amyotrophic sclerosis, from a progressive hemiplegia due to slowly increasing focal cerebral lesion involving the motor subcortex or the internal capsule, from a degenerative motor neuritis, and from a functional hemiparesis was discussed. Reasons were given why the cases did not seem to fit in exactly with any one of these diagnoses.

In addition, as has been indicated above, the diagnosis must be made from unilateral paralysis agitans. Syphilitic hemiplegia later becoming a triplegia might also have some points in common, but could usually be separated by the presence of such concomitant syphilitic affections as paralyses of the cranial nerves.

In the *Philadelphia Medical Journal*, Feb. 9, 1901, Spiller has recorded the case of a man, forty-one years old, who began, four years before coming under observation, to feel weak in the left lower limb while walking. The weakness increased, and in about a year the left upper extremity also showed signs of feebleness. He was never unconscious and never had headache or vertigo or other signs of focal intracranial disease. When the case was recorded the movements of the left lower limb were spastic, but not ataxic; the toes of the left foot were scraped along the ground and the foot turned inward. Both knee-jerks were prominent, but the left much more so than the right. Ankle clonus was obtained on the left but not on the right, and the Babinski reflex was distinct on the left, but uncertain on the right. Sensation was normal. The resistance to passive movement and the grasp of the hand were decidedly weak. The left upper limb was weaker than the right; it was also spastic, but the weakness and spasticity were less than in the lower extremity. The left upper limb was held slightly flexed at the elbow and against the body, but no contractures were present anywhere. Speech was normal, but the mouth could not be drawn up as well on the left side as on the right. The tongue went slightly to the left on protrusion. Ophthalmoscopic examination by Dr. H. F. Hansell showed white atrophy of the left optic nerve. After giving the details of his case, Spiller discusses its probable diagnosis, and concludes that the cases reported by Mills and by him have the symptoms which one should expect from unilateral lateral sclerosis, but he also called attention to the fact that other lesions might cause the same symptoms. The wasting is like that occurring in hemiplegia.

Potts (JOURNAL OF NERVOUS AND MENTAL DISEASE, Oct. 1901), under the title, "A Case of Progressive Unilateral Ascending Paralysis Probably Due to Multiple Sclerosis," has reported a case somewhat similar to the case described by the authors of this paper. The patient, a youth of nineteen, when about fifteen years old began to drag his right foot, this foot in walking showing a tendency to cross over the other. Two years after the impairment of power, diplopia appeared and remained for five months. It was discovered at about this time that the right leg was smaller than the left. Weakness in the right arm was first noticed about three and a half years after its appearance in the right leg. Examination showed that when the patient walked the right leg was rigid and the toe dragged. All movements of the leg were preserved but impaired. The right arm was distinctly weaker than the left, but was not spastic. Both the arm and leg were decidedly atrophied, as shown by careful measurements. The adductor pollicis was not so large or firm as the left. The reflexes were prompt everywhere but exaggerated on the right, the Babinski response being present. The right side of the face showed slight paresis with some tremor on voluntary effort. Paresis of the muscles of the right side of the throat was present. Nystagmus was present especially when the eyes were turned to the right. There was paresis of the left inferior rectus, and paleness of the temporal halves of the discs. All forms of sensation, including stereognosis, were normal. A slight Romberg symptom and tendency to incoördination were sometimes shown. The right hand and foot were colder to the touch than the left. Potts, while recognizing the resemblance between his case and the cases reported by the writers, holds that his case is probably one of disseminated sclerosis, and suggests that our cases might also be classed in this category, a view which we cannot accept. This diagnosis in Pott's case may be correct, although it is also probable in this and similar cases of disseminated sclerosis that the degeneration first and for a long time is chiefly confined to the pyramidal tract on one side.

Gowers, in his paper on "Abiotrophy" (*Lancet,* April 12, 1902), says truly "that the symptoms of systematic degeneration are not alone ground for the diagnosis of insular sclerosis, nor is the additional presence of nystagmus. Nystagmus on movement of the eyes is met with in many spinal degenerations of pure systemic type."

In his paper on "Chronic Progressive Hemiplegia," read at the present meeting of the American Neurological Association, Patrick describes a case which he regards as clearly indicating the progressively developing hemiplegia due to degeneration of the pyramidal tract of one side. During a recent visit to Chicago one of us (Dr. Mills) had the opportunity, through the courtesy of Dr. Patrick, of seeing this case, that of a young woman, eighteen years old, whose motor disability appeared about four years previously. The essential features of this case are the unilateral and progressive character of the paralysis which probably began first in the lower extremity, the exaggeration of the reflexes on the affected side, the absence of all forms of sensory disorder and of visceral symptoms, and the moderate wasting of the limbs and face. The reader is referred to Dr. Patrick's paper for fuller particulars of this case.

Another case of unilateral progressive ascending paralysis has been in our care at the University Hospital.

From a study of the cases presented in this paper, and in the light of the pathological findings in the case with autopsy, it may be concluded that there is a form of progressively developing hemiplegia, usually of ascending type, sometimes becoming triplegia or even quadriplegia, due to a progressive primary degeneration of the pyramidal tracts, which begins on one side and may extend to the other. This clinical picture may be produced by other lesions, but we believe that we have established the fact that primary progressive degeneration of one pyramidal tract occasionally occurs. This is a unilateral lateral sclerosis, and in this connection it is interesting to devote some space to the subject of bilateral lateral sclerosis.

In a recent address in London, Erb[2] spoke of the spastic spinal paralysis first described by him in 1875. He believes that sufficient cases have been published to show that primary bilateral sclerosis exists. The following cases he gives as indisputable, and capable of standing any criticism.

Case 1. Minkowski.[3] Clinically the picture was one of spastic spinal paralysis. Anatomically there was a nearly pure sclerosis of the crossed pyramidal tracts, with slight changes in the direct cerebellar tracts.

Case 2. Von Strümpell.[4] The patient was a man, aged sixty-three years, who had a brother suffering from the same disease. Clinically the case presented the typical picture of spastic spinal paralysis for at least twenty years. Anatomically there was a typical degeneration of the pyramidal tracts from the lumbar to the cervical region, and in addition a slight degeneration of the direct cerebellar tracts and a still more trifling degeneration of the tracts of Goll in the upper part of the spinal cord. Strümpell would, on this account, reckon the affection among the combined system diseases, but the degeneration of the pyramidal tracts was certainly the most essential lesion, and is to be regarded as primary.

Case 3. Dejerine and Sottas.[5] Clinically this was a pure case of spastic spinal paralysis of twenty-five years' duration, death occurring from pneumonia in the sixty-sixth year. Anatomically there was marked sclerosis of the pyramidal tracts from the lumbar to the cervical cord, with slight partial sclerosis of Goll's tracts in the cervical region.

Case 4. Donaggio.[6] The patient was a man aged sixty-one years. Clinically there was for two and a quarter years the typical picture of pure spastic spinal paralysis; death resulted from pneumonia. Anatomically there was pure, exclusively primary degeneration of the pyramidal tracts from the lumbar to the cervical region (a quite typical case).

Case 5. Friedmann.[7] The patient was a male, aged fifty-two years. Clinically for two years the picture was one of a spastic spinal paralysis (trace of disturbed sensation being present?). The patient suffered from apoplexy, death resulting from pneumonia. Anatomically there was classical primary degeneration of the pyramidal tracts only, there being a trace of degeneration in the direct cerebellar tracts as well as endarteritis obliterans of the basilar artery.

Case 6. Von Strümpel.[3] Clinically a picture of spastic spinal paralysis was present, death occurring after the disease had lasted for thirty-five years. Anatomically there was quite typical moderate degeneration of the pyramidal tracts from the lumbar cord to the region of the pyramids. The direct cerebellar tracts were scarcely affected and the tracts of Goll very slightly in the upper cervical region. The anterior columns and brain were entirely free (belongs to the hereditary form).

Cases 7 and 8. Bischoff.[9] The patients were two brothers who, from their eighth and tenth years respectively, suffered from spastic rigidity of the limbs, ascending slowly from the legs to the head. Intellectually development was poor, but otherwise clinically the typical picture was one of spastic spinal paralysis. Death took place from phthisis after about twenty years' duration. Anatomically there was a typical degeneration of the pyramidal tracts, extending upwards beyond the oblongata, but not farther. The direct cerebellar tracts and Gowers' bundles were all but free. Goll's tracts were extremely affected. In the gray matter of the anterior columns there was atrophy of the ganglion cells (evidently occurring towards the end of life). The author calls the change in the spinal cord primary tract degeneration (quite analogous to the condition found by Strümpell), and holds that thereby the existence of an infantile form of hereditary spastic spinal paralysis has been proved.

Case 9. Ida Democh.[10] The case is clinically and anatomically somewhat complicated; clinically there was a typical picture of spastic spinal paralysis, combined with signs of chronic alcoholism, pains, tremors, etc. Anatomically there were primary degeneration of the tracts of Goll and congenital hydromyelia in the lumbar and dorsal cord; the direct cerebellar tracts were free; nevertheless, as proved by the author, this can be termed a case of primary degeneration of the pyramidal tracts in spastic spinal paralysis.

Case 10. A case clinically typical of spastic spinal paralysis described by Kühn, in which von Strümpell made the microscopical examination of the spinal cord. He found an essentially characteristic primary degeneration of the lateral columns.

Erb regarded the sclerosis of the direct cerebellar tracts and the slight degeneration of the columns of Goll in some of these cases as of secondary importance.

He thinks that the case described by Strümpell[11] in 1894 as a primary isolated system degeneration of both pyramidal tracts, showing only slight transitions to amyotrophic lateral sclerosis, might come under this head.

To this record we would add the case reported by us in this paper, and also the case reported by one of us (Spiller[12]) in which there was primary degeneration of the pyramidal tracts with comparatively slight alteration of the nerve cells of the anterior horns of the spinal cord.

Note—Since this paper was read, another communication by Erb on spastic spinal paralysis has been published in the *Deutsche Zeitschrift für Nervenheilkunde,* Vol. 23, Nos. 5 and 6. He refers to the ten cases mentioned in his previous paper, and includes the well known case of Morgan and Dreschfeld.

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[2] Erb, Lancet, 1902, p. 970.

[3] Minkowski, Deutsches Archiv f. klin. Med., Vol. 34, 1884.

[4] Von Strümpell, Archiv f. Psych., 17, 1886.

[5] Dejerine and Sottas, Archives de Physiologie Normale et Pathologique, 1896, p. 630.

[6] Donaggio, Rivista sperimentale di freniatria, Vol. 23, 1897.

[7] Friedmann, Deutsche Zeitschr. f. Nervenheilk., Vol. 16, 1899.

[8] Von Strümpell, Neur. Centralbl., 1901, p. 530.

[9] Bischoff, Jahrbücher f. Psych. und Neur., Vol. 22, 1902.

[10] Ida Democh, Archiv. f. Psych., Vol. 33, 1900.

[11] Von Strümpell, Deut. Zeitschr. f. Nerv., Vol. 5, p. 225.

[12] Spiller, Journal of Nervous and Mental Disease, May, 1902.